

Lacrimal duct problems

1. Dacryocystitis

It is infection of the lacrimal sac

Features:

- 1) watering eye (epiphora)
- 2) swelling and erythema at the inner canthus of the eye

Management:

- 1) With systemic antibiotics.
- 2) Intravenous antibiotics are indicated if there is associated periorbital cellulitis

2. Congenital lacrimal duct obstruction:

- It affects around 5-10% of newborns.
- It is bilateral in around 20% of cases

Features:

- 1) watering eye (even if not crying)
- 2) secondary infection may occur
- 3) Symptoms resolve in 99% of cases by 12 months of age

VISIT...

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Blepharitis

- Blepharitis is inflammation of the eyelid margins.
- It may be due to either:
 - 1) **Meibomian gland dysfunction** (common, posterior blepharitis) or
 - 2) **Seborrhoeic dermatitis/Staphylococcal infection** (less common, anterior blepharitis).
- Blepharitis is also more common in patients with Acne rosacea
- The meibomian glands secrete oil on to the eye surface to prevent rapid evaporation of the tear film. Any problem affecting the meibomian glands (as in blepharitis) can hence cause drying of the eyes which in turn leads to irritation

Features:

- 1) Symptoms are usually bilateral
- 2) Grittiness and discomfort, particularly around the eyelid margins
- 3) Eyes may be sticky in the morning
- 4) Eyelid margins may be red.
- 5) Swollen eyelids may be seen in staphylococcal blepharitis
- 6) Styes and chalazions are more common in patients with blepharitis
- 7) Secondary conjunctivitis may occur

Management

- 1) softening of the lid margin using **hot compresses twice a day**
- 2) **mechanical removal of the debris** from lid margins: - cotton wool buds dipped in a mixture of **cooled boiled water** and **baby shampoo** is often used*
- 3) **artificial tears** may be given for symptom relief in people with dry eyes or an abnormal tear film

*an alternative is **sodium bicarbonate**, a teaspoonful in a cup of cooled water that has recently been boiled

Herpes Simplex Keratitis

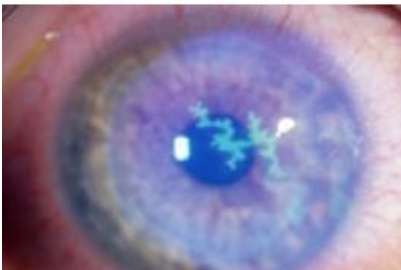
Herpes simplex keratitis most commonly presents with a **dendritic corneal ulcer**

Features:

- 1) red, painful eye
- 2) photophobia
- 3) epiphora
- 4) visual acuity may be decreased
- 5) fluorescein staining may show an epithelial ulcer, dendritic pattern of staining

Management:

- 1) Immediate referral to an ophthalmologist
- 2) Topical aciclovir



Herpes zoster ophthalmicus

- Herpes zoster ophthalmicus (HZO) describes the reactivation of the varicella zoster virus in the area supplied by the ophthalmic division of the trigeminal nerve.
- It accounts for around 10% of case of shingles.

Features:

- 1) vesicular rash around the eye, which may or may not involve the actual eye itself
- 2) **Hutchinson's sign**: rash on the tip or side of the nose. Indicates nasociliary involvement and is a strong risk factor for ocular involvement

Management:

- 1) **Oral antiviral** treatment for **7-10 days ideally started within 72 hours**.
Topical antiviral treatment is **not given in HZO**
- 2) **Oral corticosteroids** may reduce the duration of pain but do not reduce the incidence of post-herpetic neuralgia
- 3) Ocular involvement requires urgent ophthalmology review

Complications:

- 1) ocular: conjunctivitis, keratitis, episcleritis, anterior uveitis
- 2) ptosis
- 3) post-herpetic neuralgia

Band keratopathy

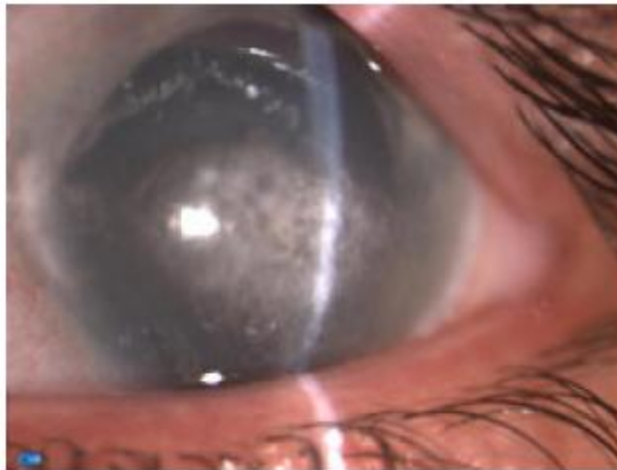
- **It** is a corneal disease derived from the appearance of calcium on the central cornea caused by calcium deposition in Bowman's layer
- This is an example of metastatic calcification which by definition, occurs in the presence of hypercalcaemia

Symptoms include pain and decreased visual acuity.

Treatment:

The calcium can be scraped off the cornea or removed with a laser.

- ✓ This can restore sight, but it can take a number of months for normal vision to return as the cornea will be damaged during the operation.
- ✓ This cannot be repeated too many times as it would make the cornea thinner and thinner



Cataracts

Majority:

- 1) age related
- 2) UV light

Systemic:

- 1) Diabetes mellitus
- 2) Steroids
- 3) Infection (congenital rubella)
- 4) Metabolic (hypocalcaemia, galactosaemia)
- 5) Myotonic dystrophy, down's syndrome

Ocular:

- 1) trauma
- 2) uveitis
- 3) high myopia
- 4) topical steroids

Classification:

- 1) **Nuclear:** change lens refractive index, common in old age
- 2) **Polar:** localized, commonly inherited, lie in the visual axis
- 3) **Subcapsular:** due to steroid use, just deep to the lens capsule, in the visual axis
- 4) **Dot opacities:** common in normal lenses, also seen in diabetes and myotonic dystrophy

Lens dislocation

Causes:

- 1) Marfan's syndrome: upwards
- 2) homocystinuria: downwards
- 3) Ehlers-Danlos syndrome
- 4) trauma
- 5) uveal tumours
- 6) autosomal recessive ectopia lentis

Anterior uveitis

Anterior uveitis is one of the important differentials of a red eye. It is also referred to as iritis.

Features:

- 1) acute onset
- 2) ocular discomfort & pain (may increase with use)
- 3) pupil may be irregular and small
- 4) photophobia (often intense)
- 5) blurred vision
- 6) red eyes
- 7) lacrimation
- 8) ciliary flush
- 9) visual acuity initially normal → impaired

Associated conditions:

- 1) ankylosing spondylitis
- 2) reactive arthritis
- 3) ulcerative colitis, Crohn's disease
- 4) Behcet's disease

Management:

- 1) urgent review by ophthalmology
- 2) cycloplegics (dilates the pupil which helps to relieve pain and photophobia) e.g. **Atropine, cyclopentolate**
- 3) **steroid eye drops**

Acute angle closure glaucoma

- Glaucoma is a group disorders characterised by optic neuropathy due, in the majority of patients, to raised intraocular pressure (IOP).
- It is now recognised that a minority of patients with raised IOP do not have glaucoma and vice versa
- In acute angle closure glaucoma (AACG) there is a rise in IOP **secondary to impairment of aqueous outflow**.

Factors predisposing to AACG include:

- 1) hypermetropia (long-sightedness)
- 2) pupillary dilatation
- 3) lens growth associated with age

Features:

- 1) severe pain: may be ocular or headache
- 2) decreased visual acuity
- 3) symptoms worse with mydriasis (e.g. watching TV in a dark room)
- 4) hard, red eye
- 5) haloes around lights
- 6) semi-dilated non-reacting pupil
- 7) corneal oedema results in dull or hazy cornea
- 8) systemic upset may be seen, such as nausea and vomiting and even abdominal pain

Management:

- 1) urgent referral to an ophthalmologist
- 2) management options include:
 - A) reducing aqueous secretions with **acetazolamide** and
 - B) inducing pupillary constriction with **topical pilocarpine**

Primary open-angle glaucoma

- Glaucoma is a group disorders characterised by optic neuropathy due, in the majority of patients, to raised intraocular pressure (IOP).
- It is now recognised that a minority of patients with raised IOP do not have glaucoma and vice versa
- Primary open-angle glaucoma (POAG) also referred to as **chronic simple glaucoma**
- It is present in around **2% of people older than 40 years**.
- Other than age, risk factors include:
 - 1) family history
 - 2) black patients
 - 3) myopia
 - 4) hypertension
 - 5) diabetes mellitus

POAG may present **insidiously** and for this reason is often **detected during routine optometry appointments**.

Features may include

- 1) peripheral visual field loss - nasal scotomas progressing to 'tunnel vision'
- 2) decreased visual acuity
- 3) optic disc cupping

Management:

- 1) The majority of patients with primary open-angle glaucoma are managed with **eye drops**.
- 2) These aim to lower intra-ocular pressure which in turn has been shown to prevent progressive loss of visual field.
- 3) Surgery in the form of a **trabeculectomy** may be considered in refractory cases.

Medication	Mode of action	Notes
Prostaglandin analogues (e.g. Latanoprost)	Increases uveoscleral outflow	• Once daily administration • Adverse effects include brown pigmentation of the iris
Miotics (e.g. pilocarpine, a muscarinic receptor agonist)	Increases uveoscleral outflow	Adverse effects included a constricted pupil, headache and blurred vision
Beta-blockers (e.g. Timolol)	Reduces aqueous production	• Should be avoided in asthmatics and patients with heart block
Carbonic anhydrase inhibitors (Dorzolamide)	Reduces aqueous production	• Systemic absorption may cause sulphonamide-like reactions
Sympathomimetics (e.g. brimonidine, an alpha2-adrenoceptor agonist)	1) Reduces aqueous production and 2) increases outflow	• Avoid if taking MAOI or tricyclic antidepressants • Adverse effects include hyperaemia

Age related macular degeneration

- Age related macular degeneration is the most common cause of blindness in the UK.
- Degeneration of the central retina (macula) is the key feature with changes usually bilateral
- Traditionally two forms of macular degeneration are seen:

A) Dry (geographic atrophy) macular degeneration:

Characterised by drusen (yellow round spots in Bruch's membrane)

B) Wet (exudative, neovascular) macular degeneration:

- ✓ Characterised by choroidal neovascularisation.
- ✓ Leakage of serous fluid and blood can subsequently result in a rapid loss of vision.
- ✓ Carries worst prognosis

Recently there has been a move to a more updated classification:

A) Early age related macular degeneration (non-exudative, age related maculopathy):
drusen and alterations to the retinal pigment epithelium (RPE)

B) Late age related macular degeneration (neovascularisation, exudative)

Risk factors:

- 1) age: most patients are over 60 years of age
- 2) female sex
- 3) family history
- 4) more common in Caucasians
- 5) smoking
- 6) high cumulative sunlight exposure

Features:

- 1) reduced visual acuity: 'blurred', 'distorted' vision, central vision is affected first
- 2) central scotomas
- 3) fundoscopy: drusen, pigmentary changes

Investigation and diagnosis:

- 1) **Optical coherence tomography:** provide cross sectional views of the macula
- 2) if neovascularisation is present **fluorescein angiography** is performed

General management:

- 1) stop smoking
- 2) High dose of beta-carotene, vitamins C and E, and zinc may help to slow down visual loss for patients with established macular degeneration.
- 3) **Supplements should be avoided in smokers** due to an **increased risk of lung cancer**

A) Dry macular degeneration: no current medical treatments

B) Wet macular degeneration:

- 1) Photocoagulation
- 2) Photodynamic therapy
- 3) Anti-vascular endothelial growth factor (anti-VEGF) treatments: intravitreal ranibizumab



Normal Vision



*Same view with age related
Macular Degeneration*

Angioid Retinal Streaks

- Angioid retinal streaks are seen on fundoscopy as irregular dark red streaks radiating from the optic nerve head.
- They are caused by degeneration, calcification and breaks in Bruch's membrane.

Causes: ASPEP

- 1) Acromegaly
- 2) Sickle-cell anaemia
- 3) Paget's disease
- 4) Ehler-Danlos syndrome
- 5) Pseudoxanthoma elasticum



Choroidoretinitis

Causes

- 1) syphilis
- 2) cytomegalovirus
- 3) toxoplasmosis
- 4) sarcoidosis
- 5) tuberculosis

Diabetic Retinopathy

- Diabetic retinopathy is the most common cause of blindness in adults aged 35-65 years-old.
- Hyperglycaemia is thought to cause increased retinal blood flow and abnormal metabolism in the retinal vessel walls.
- This precipitates damage to endothelial cells and pericytes.
- **Endothelial dysfunction leads to increased vascular permeability** which causes the characteristic **exudates** seen on fundoscopy.
- **Pericyte dysfunction** predisposes to the formation of **microaneurysms**.
- **Neovascularization** is thought to be caused by the production of **growth factors** in response to **retinal ischaemia**.
- In exams you are most likely to be asked about the characteristic features of the various stages/types of diabetic retinopathy.
- Recently a new classification system has been proposed, dividing patients into those with non-proliferative diabetic retinopathy (NPDR) and those with proliferative retinopathy (PDR):

Traditional classification	New classification
Background retinopathy: 1) microaneurysms (dots) 2) blot haemorrhages (≤ 3) 3) hard exudates	Mild NPDR • 1 or more microaneurysm
Pre-proliferative retinopathy: 1) cotton wool spots (soft exudates; ischaemic nerve fibres) 2) > 3 blot haemorrhages 3) venous beading/looping 4) deep/dark cluster haemorrhages more common in Type I DM, treat with laser photocoagulation	Moderate NPDR 1) microaneurysms 2) blot haemorrhages 3) hard exudates 4) cotton wool spots, venous beading/looping and intraretinal microvascular abnormalities (IRMA) less severe than in severe NPDR
	Severe NPDR 1) blot haemorrhages and microaneurysms in 4 quadrants 2) venous beading in at least 2 quadrants 3) IRMA in at least 1 quadrant

Proliferative retinopathy:

- 1) retinal neovascularisation - may lead to vitreous haemorrhage
- 2) fibrous tissue forming anterior to retinal disc
- 3) more common in Type I DM, 50% blind in 5 years

Maculopathy:

- 1) based on location rather than severity, anything is potentially serious
- 2) hard exudates and other 'background' changes on macula
- 3) check visual acuity
- 4) more common in Type II DM

Tunnel vision

Tunnel vision is the concentric diminution of the visual fields

Causes:

- 1) papilloedema
- 2) glaucoma
- 3) retinitis pigmentosa
- 4) choroidoretinitis
- 5) optic atrophy secondary to tabes dorsalis
- 6) hysteria

Retinitis pigmentosa

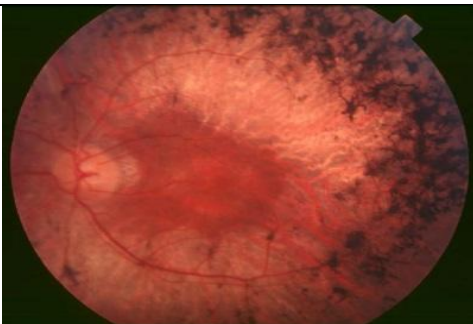
Retinitis pigmentosa primarily affects the peripheral retina resulting in funnel vision

Features:

- 1) night blindness is often the initial sign
- 2) funnel vision (the preferred term for tunnel vision)
- 3) fundoscopy: black bone spicule-shaped pigmentation in the peripheral retina, mottling of the retinal pigment epithelium

Associated diseases:

- 1) Refsum disease: cerebellar ataxia, peripheral neuropathy, deafness, ichthyosis
- 2) Usher syndrome
- 3) abetalipoproteinemia
- 4) Lawrence-Moon-Biedl syndrome
- 5) Kearns-Sayre syndrome
- 6) Alport's syndrome



Fundus showing changes secondary to retinitis pigmentosa



Tunnel vision

Congenital Hypertrophy of the Retinal Pigment Epithelium (CHRPE):

A) Typical CHRPE:

- Gray or black depigmented lacunae
- Found in 1 quadrant of eye
- Do not affect vision

B) Atypical CHRPE:

- White fish tail shaped bilaterally
- Affect the vision when there are > 4 in each eye
- Associated with Adenosis polyposis and **Gardner's syndrome** → do colonoscopy.

Papilloedema

The following features may be observed during fundoscopy:

- 1) venous engorgement: usually the first sign
- 2) loss of venous pulsation: although many normal patients do not have normal pulsation
- 3) blurring of the optic disc margin
- 4) elevation of optic disc
- 5) loss of the optic cup
- 6) Paton's lines: concentric/radial retinal lines cascading from the optic disc

Causes of papilloedema:

- 1) space-occupying lesion: neoplastic, vascular
- 2) malignant hypertension
- 3) idiopathic intracranial hypertension
- 4) hydrocephalus
- 5) hypercapnia

Rare causes include:

- 1) hypoparathyroidism and
- 2) hypocalcaemia
- 3) vitamin A toxicity

Optic neuritis

Causes:

- 1) Multiple sclerosis
- 2) Diabetes
- 3) Syphilis

Features:

- 1) unilateral decrease in visual acuity over hours or days
- 2) poor discrimination of colours, 'red desaturation'
- 3) pain worse on eye movement
- 4) relative afferent pupillary defect
- 5) central scotoma

Management:

- 1) **high-dose steroids**
- 2) **recovery usually takes 4-6 weeks**

Prognosis:

- MRI: if > 3 white-matter lesions, 5-year risk of developing multiple sclerosis is c. 50%



Unilateral decrease in visual acuity over hours or days

Retrobulbar Neuritis

Inflammation behind the optic nerve head, the optic disc is normal.
Patient sees nothing, Doctor sees nothing

Features:

- 1) Visual acuity loss
- 2) Afferent pupillary defect during swinging flashing light
- 3) Color vision will be reduced (red looks pallor)

Optic atrophy

- Optic atrophy is seen as pale, well demarcated disc on fundoscopy.
- It is usually bilateral and causes a gradual loss of vision*.
- Causes may be acquired or congenital

Acquired causes:

- 1) multiple sclerosis
- 2) papilloedema (longstanding)
- 3) raised intraocular pressure (e.g. glaucoma, tumour)
- 4) retinal damage (e.g. choroiditis, retinitis pigmentosa)
- 5) ischaemia
- 6) toxins: tobacco amblyopia, quinine, methanol, arsenic, lead
- 7) nutritional: vitamin B1, B2, B6 and B12 deficiency

Congenital causes:

- 1) Friedreich's ataxia
- 2) mitochondrial disorders e.g. Leber's optic atrophy
- 3) DIDMOAD - the association of cranial Diabetes Insipidus, Diabetes Mellitus, Optic Atrophy and Deafness (also known as Wolfram's syndrome)

*strictly speaking optic atrophy is a descriptive term, it is the optic neuropathy that results in visual loss

Relative afferent pupillary defect

- Also known as the Marcus-Gunn pupil,
- A relative afferent pupillary defect is found by the 'swinging light test'.
- It is caused by a lesion anterior to the optic chiasm i.e. optic nerve or retina

Causes:

- 1) retina: detachment
- 2) optic nerve: optic neuritis e.g. multiple sclerosis

Pathway of pupillary light reflex:

- afferent: retina → optic nerve → lateral geniculate body → midbrain
- efferent: Edinger-Westphal nucleus (midbrain) → oculomotor nerve

Temporal arteritis

- Temporal arteritis is large vessel vasculitis which overlaps with polymyalgia rheumatica (PMR).
- Histology shows changes which characteristically 'skips' certain sections of affected artery whilst damaging others.

Features:

- 1) typically patient > 60 years old
- 2) usually rapid onset (e.g. < 1 month)
- 3) headache (found in 85%)
- 4) jaw claudication (65%)
- 5) visual disturbances secondary to anterior ischemic optic neuropathy
- 6) tender, palpable temporal artery
- 7) features of PMR: aching, morning stiffness in proximal limb muscles (not weakness)
- 8) also lethargy, depression, low-grade fever, anorexia, night sweats

Investigations:

- 1) Raised inflammatory markers: ESR > 50 mm/hr (note ESR < 30 in 10% of patients). CRP may also be elevated
- 2) temporal artery biopsy: skip lesions may be present
- 3) Note creatine kinase and EMG normal

Treatment:

- 1) high-dose prednisolone - there should be a dramatic response, if not the diagnosis should be reconsidered
- 2) urgent ophthalmology review. Patients with visual symptoms should be seen the same-day by an ophthalmologist. Visual damage is often irreversible

Red eye

- There are many possible causes of a red eye.
 - It is important to be able to recognise the causes which require urgent referral to an ophthalmologist.
 - Below is a brief summary of the key distinguishing features:
1. **Acute angle closure glaucoma:**
 - 1) severe pain (may be ocular or headache)
 - 2) decreased visual acuity, patient sees haloes
 - 3) semi-dilated pupil
 - 4) hazy cornea
 2. **Anterior uveitis:**
 - 1) Acute onset
 - 2) Pain
 - 3) Blurred vision and photophobia
 - 4) Small, fixed oval pupil, ciliary flush
 3. **Scleritis:**
 - 1) severe pain (may be worse on movement) and tenderness
 - 2) may be underlying autoimmune disease e.g. rheumatoid arthritis
 4. **Conjunctivitis:**
 - 1) purulent discharge if bacterial,
 - 2) clear discharge if viral
 5. **Subconjunctival haemorrhage:**
 - history of trauma or coughing bouts

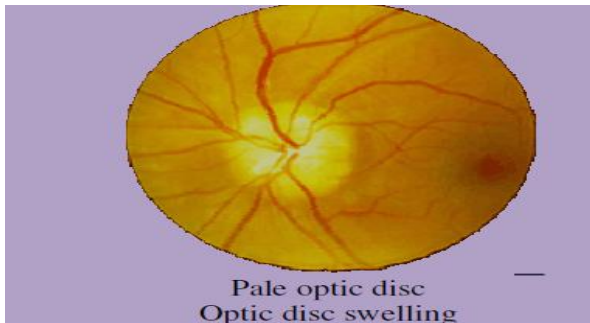
Sudden Painless Loss Of Vision

The most common causes of a sudden painless loss of vision are as follows:

- 1) ischaemic optic neuropathy (e.g. temporal arteritis or atherosclerosis)
- 2) occlusion of central retinal vein
- 3) occlusion of central retinal artery
- 4) vitreous haemorrhage
- 5) retinal detachment

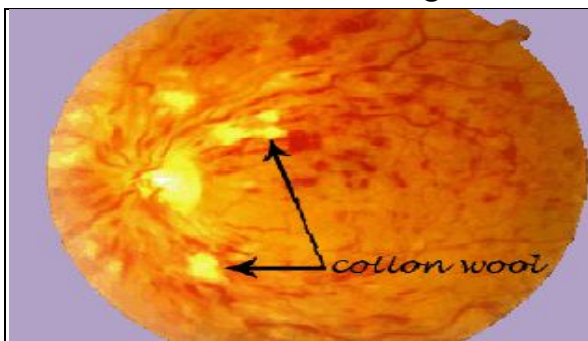
1. Ischaemic optic neuropathy:

- 1) may be due to:
 - A. arteritis (e.g. temporal arteritis) or
 - B. atherosclerosis (e.g. hypertensive, diabetic older patient)
- 2) due to occlusion of the short posterior ciliary arteries, causing damage to the optic nerve
- 3) altitudinal field defects are seen



2. Central retinal vein occlusion

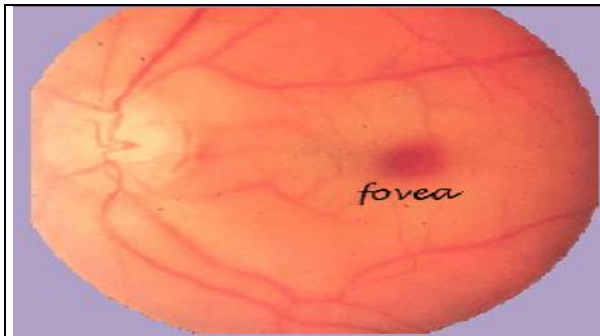
- 1) incidence increases with age, more common than arterial occlusion
- 2) causes: glaucoma, polycythaemia, hypertension
- 3) severe retinal haemorrhages are usually seen on fundoscopy



- Widespread retinal hemorrhages in all 4 quadrants, which vary in appearance from a small-scattered retinal hemorrhages to marked confluent hemorrhages
- Marked dilated and tortuous retinal vessels
- Cotton-wool spots
- Optic disc edema, macular edema, and retinal thickening
- Vitreous hemorrhages may be present

3. Central retinal artery occlusion

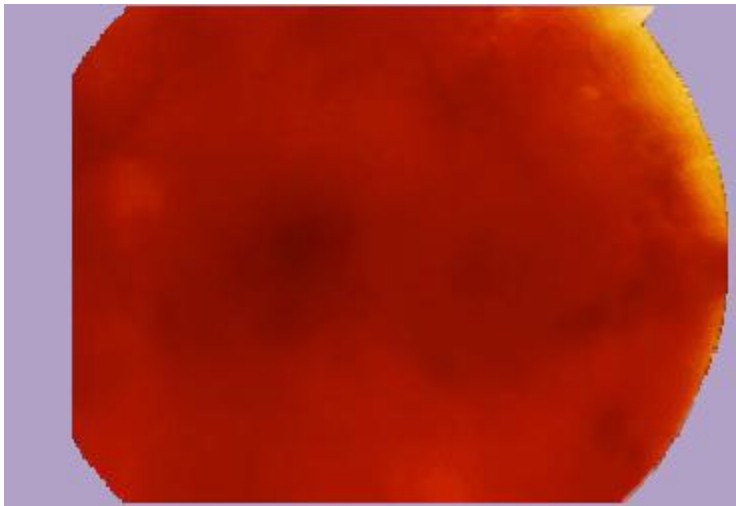
- 1) due to thromboembolism (from atherosclerosis) or arteritis (e.g. temporal arteritis)
- 2) features include afferent pupillary defect, 'cherry red' spot on a pale retina



- Diffuse edema makes the retina and arteries look pale.
- Perfused underlying tissues show through the thin fovea giving a classic cherry-red spot appearance.

4. Vitreous haemorrhage:

- 1) causes: diabetes, bleeding disorders
- 2) features may include sudden visual loss, dark spots



5. Retinal detachment:

- features of vitreous detachment, which may precede retinal detachment, include flashes of light or floaters (see below)

Differentiating posterior vitreous detachment, retinal detachment and vitreous haemorrhage

Posterior vitreous detachment	Retinal detachment	Vitreous haemorrhage
1) Flashes of light (photopsia) in the peripheral field of vision 2) Floaters, often on the temporal side of the central vision	1) Dense shadow that starts peripherally progresses towards the central vision 2) A veil or curtain over the field of vision 3) Straight lines appear curved 4) Central visual loss	1) Large bleeds cause sudden visual loss 2) Moderate bleeds may be described as numerous dark spots 3) Small bleeds may cause floaters

Mydriasis

Causes of mydriasis (large pupil):

- 1) third nerve palsy
- 2) Holmes-Adie pupil
- 3) traumatic iridoplegia
- 4) pheochromocytoma
- 5) congenital

Drug causes of mydriasis

- 1) topical mydriatics: tropicamide, atropine
- 2) sympathomimetic drugs: amphetamines, cocaine
- 3) anticholinergic drugs: tricyclic antidepressants

Miosis

Causes of miosis (small pupil)

- 1) Horner's syndrome
- 2) Argyll-Robertson pupil
- 3) senile miosis
- 4) pontine haemorrhage
- 5) congenital

Drugs causes:

- 1) opiates
- 2) parasympathomimetics: pilocarpine
- 3) organophosphate toxicity

Ptosis

Ptosis may be unilateral or bilateral

Causes of bilateral ptosis:

- 1) myotonic dystrophy
- 2) myasthenia gravis
- 3) syphilis
- 4) congenital

Causes of unilateral ptosis:, as above plus:

- 1) third nerve palsy
- 2) Horner's

Horner's syndrome

Features:

- 1) ptosis
- 2) miosis (small pupil)
- 3) anhydrosis (loss of sweating one side)
- 4) enophthalmos* (sunken eye)

*in reality the appearance is due to a narrow palpebral aperture rather than true enophthalmos

Distinguishing between causes

- 1) heterochromia (difference in iris colour) is seen in congenital Horner's
- 2) anhydrosis: see below

Central lesions	Pre-ganglionic lesions	Post-ganglionic lesions
Anhydrosis of the face, arm and trunk	Anhydrosis of the face	No anhydrosis
Stroke Syringomyelia Multiple sclerosis Tumour Encephalitis	Pancoast's tumour Thyroidectomy Trauma Cervical rib	Carotid artery dissection Carotid aneurysm Cavernous sinus thrombosis Cluster headache

Holmes-Adie pupil (Myotonic pupil)

- It is a benign condition most commonly seen in young women
- it is common and usually **unilateral** (80% of cases)
- May occur after an episode of zoster infection.
- **This is a dilated, often irregular pupil;**
- once the pupil has constricted it remains small for an abnormally long time
- Slowly reactive to accommodation but very poorly (if at all) to light.
- This is due to denervation in the ciliary ganglion, of unknown cause, and has no other pathological significance.
- At the beginning of the condition the pupil is large, but over time becomes small and poorly reactive.
- In adults it tends to be a benign condition and is simply observed, however infants are usually referred because of an association with familial dystonias

Diagnosis:

- 1) Slit lamp examination may reveal small worm like contractions of the iris, but
- 2) **pilocarpine eye drops:**
 - ✓ The usual diagnostic test
 - ✓ Use weak pilocarpine eye drops, which induce vigorous pupil contraction on the affected side, but only weak contraction of the pupil on the unaffected side.

Holmes-Adie syndrome

- association of Holmes-Adie pupil with absent ankle/knee reflexes

Argyll Robertson pupil (**Accommodate but not react; prostitute pupil**)

- Argyll-Robertson pupil is one of the classic pupillary syndromes. (Now rarely seen in clinical practice)
- It is sometimes seen in neurosyphilis and is often said to be the prostitute's pupil - accommodates but doesn't react!
- Another mnemonic used for the Argyll-Robertson Pupil (ARP) is Accommodation Reflex Present (ARP) but Pupillary Reflex Absent (PRA)

Features:

- 1) **A small, irregular pupil**
- 2) no response to light but there is a response to accommodate

Causes:

- 1) syphilis (Once Considered diagnostic of neurosyphilis)
- 2) diabetes mellitus (It is now only occasionally seen in diabetes or MS)
- 3) The lesion is in the brainstem surrounding the aqueduct of Sylvius.

Trochlear Nerve Palsy: cause torsional diplopia, Torsion is a normal response to tilting the head sideways. The eyes automatically rotate in an equal and opposite direction, so that the orientation of the environment remains unchanged – vertical things remain vertical.

Rheumatoid arthritis: ocular manifestations

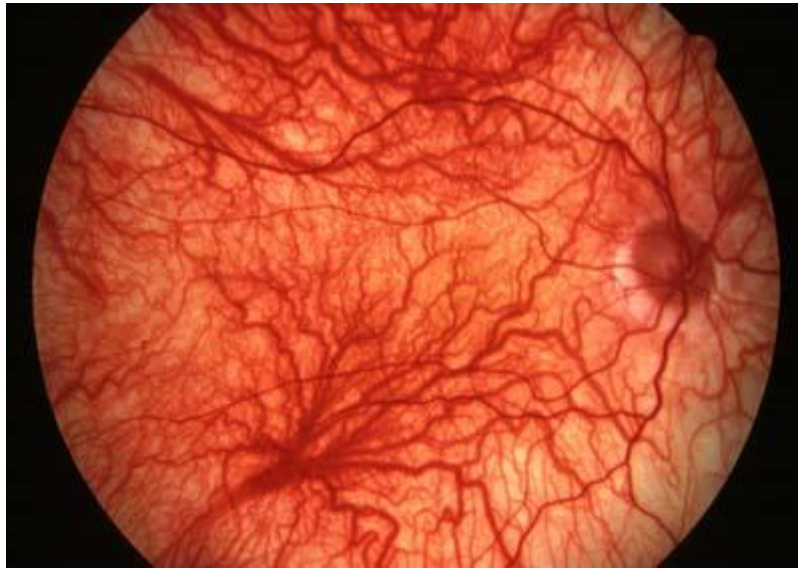
Ocular manifestations of rheumatoid arthritis are common, with 25% of patients having eye problems

Ocular manifestations:

- 1) keratoconjunctivitis sicca (most common)
- 2) episcleritis (erythema)
- 3) scleritis (erythema and pain)
- 4) corneal ulceration
- 5) keratitis

Iatrogenic:

- 1) steroid-induced cataracts
- 2) chloroquine retinopathy



This is an albino fundus. There is no retinal pigmentation and all the blood vessels can be clearly seen.

Nystagmus and photophobia are common findings in albinos.

Albinism represents a group of inherited abnormalities of melanin synthesis characterised by a congenital reduction or absence of melanin pigment, in association with specific developmental changes in the optic system resulting from the hypopigmentation.

Oculocutaneous albinism (OCA) involves two regions of the body:

1. The skin and hair
2. The optic system including the eye and the optic nerves.

Ocular albinism (OA) has the same changes in the optic system by reducing mainly the pigment in the retinal pigment epithelium of the eye, usually with no clinical difference in the colour of the skin and hair.

The albino macula is always hypoplastic, and the patient has reduced acuity and pendular nystagmus. Strabismus is also common.

Oculocutaneous albinism

Tyrosinase-related oculocutaneous albinism (OCA1)

This is an autosomal recessive disorder characterised by absence of pigment in hair, skin, and eyes, and does not vary with race or age. Severe nystagmus, photophobia, and reduced visual acuity are common features.

OCA1 is divided into two types:

1. Type IA, characterised by complete lack of tyrosinase activity due to production of an inactive enzyme.
2. Type IB (also called yellow OCA), characterised by reduced activity of tyrosinase.

P-gene related oculocutaneous albinism (OCA2)

OCA2 is the most common type of albinism, and is especially frequent among African-Americans and Africans. The estimated frequency of OCA2 in the African-

American population is 1 in 10,000, which contrasts with a frequency of 1 in 36,000 in Caucasians.

Chediak-Higashi syndrome

This is autosomal recessive. Patients may have a silvery sheen to their skin, and blue to brown irises. Patients have an increased susceptibility to infection, hepatosplenomegaly, lymphadenopathy and a predisposition to development of a lymphoma-like condition.

Hermansky-Pudlak syndrome

This is autosomal recessive. It is associated with the absence of platelet-dense bodies, resulting in a loss of secondary AGGREGATION of platelets after stimulation, and a predisposition to bruising and bleeding which can be severe. There is a higher frequency in Puerto Rico.

Ocular albinism

X-linked ocular albinism type 1 (OA1)

Although this type of albinism is categorised as a type of ocular albinism, the melanocytes in the skin and hair follicles are also involved. Individuals with OA1 have normal-looking cutaneous pigment, variable iris pigment, and reduced or absent retinal pigment associated with foveal hypoplasia and optic tract misrouting. The OA1 gene maps to the X chromosome at Xp22.3.

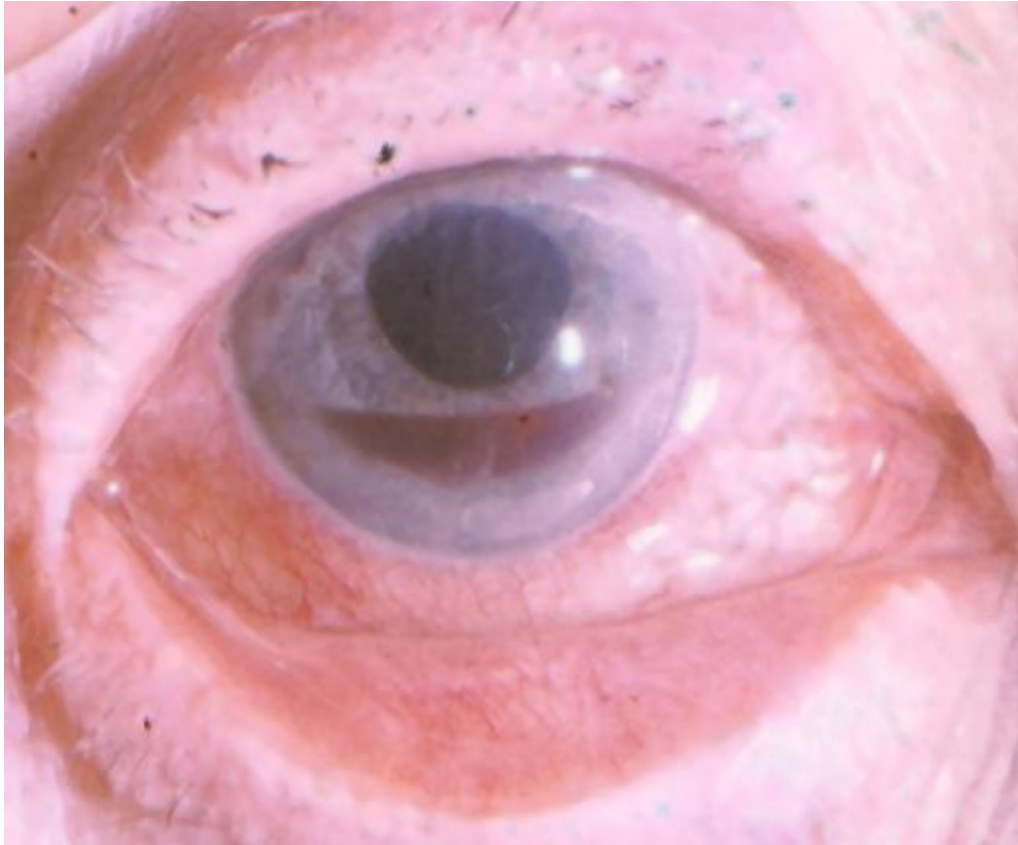
Other options listed in the question.

Abetalipoproteinaemia is associated with retinitis pigmentosa.

Chronic granulomatous disease presents with recurrent infections due to an inability to generate the oxidative burst necessary for phagocyte killing; it is not associated with any fundal abnormality.

Cystic fibrosis is associated with recurrent lower respiratory tract infections but not with fundal abnormality.

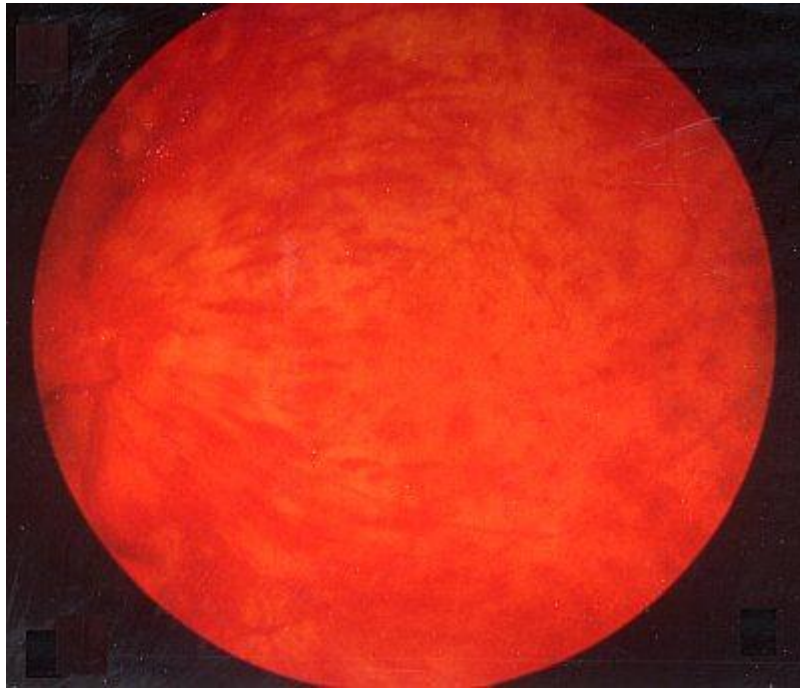
Type 1 DM is associated with infections and eye disease - though diabetic retinopathy does not have this appearance.



The slide shows hyphaema: blood in the anterior chamber.

It is usually caused by trauma - often small objects (champagne corks, squash balls) hitting the eye.

Aspiration may be required to prevent loss of vision.



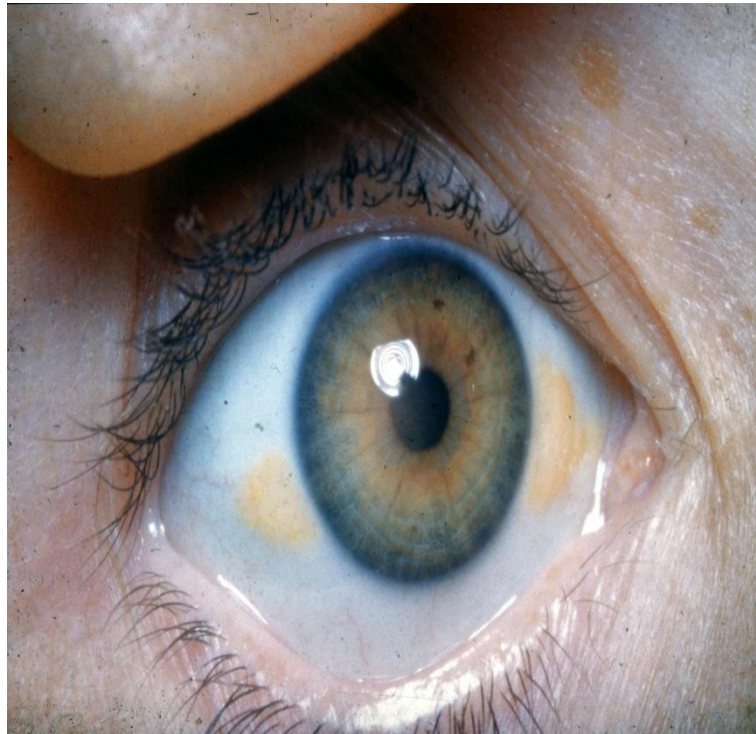
The slide shows the typical appearance of central retinal vein occlusion. Central retinal vein occlusion is most common in elderly patients, secondary to:

- glaucoma
- diabetes mellitus
- hypertension
- increased blood viscosity
- high haematocrit
- optic disc edema
- hypercoagulable states
- vasculitis
- retrobulbar compression by tumors, or
- Grave's ophthalmopathy.

In the young person it can be idiopathic or the result of retinal phlebitis.

The presentation of central retinal vein occlusion has been described as follows¹:

"Patients usually present with painless loss of vision and are found to have diffuse retinal hemorrhages in all four quadrants of the retina as well as dilated, tortuous veins, cotton-wool spots, disc edema, optociliary shunt vessels and neovessels might also be present."



The slide shows yellow papules (pingueculae) in the cornea; these are characteristic of Gaucher disease.

Gaucher disease is inherited as an autosomal recessive disease. The disease is caused by a deficiency of the enzyme glucocerebrosidase, essential for the metabolism of glycolipids.

There are three types of Gaucher disease:

- Type 1: Chronic non-neuropathic; adult Gaucher disease
- Type 2: Acute neuropathic; infantile Gaucher disease
- Type 3: Subacute neuropathic; juvenile Gaucher disease

Patients with all types of disease have hepatosplenomegaly and large glucocerebrosidase-rich cells (Gaucher cells) infiltrating the bone marrow.

Type 2, infantile Gaucher disease, carries the worst prognosis, with children seldom surviving beyond 2 years.

Type 1 disease is the commonest, usually presenting in childhood with hepatosplenomegaly, but not uncommonly in middle or old age.

Bone marrow replacement and hypersplenism result in anaemia and thrombocytopenia.

Pathological bone fractures and avascular necrosis of the femoral heads are not uncommon.

Bony disease may be confined to the distal ends of the femurs, with formation of characteristic 'Erlenmeyer flask' shaped cysts.

The skin may show a grey-brown discolouration, especially around the forehead, hands and pre-tibial regions. Characteristic yellow or yellow-brown papules (pingueculae) develop at the sclerocorneal junctions.



Loss of night vision and peripheral vision are classic features of retinitis pigmentosa.
The fundi shows the characteristic 'bone spicule' areas of pigmentation in the periphery of the retina.



The fundus shows small pale dots over the macular area typical of drusen.
This is macular degeneration and one of the commonest causes of blindness.